The gallbladder, cystic duct, tumour, part of the CHD and the supraduodenal portion of CBD were excised en masse. Continuity was restored by hepatico-jejunostomy.

Histopathology revealed a poorly differentiated adenocarcinoma of the cystic duct. She had an uneventful recovery and was discharged on the 14th postoperative day. Nine months later, she remained well with normal liver function. Repeat ultrasound showed no evidence of recurrence or metastasis.

Discussion

Primary carcinoma of the extrahepatic biliary tree is rare, an incidence of 0.14% was found in 46 593 autopsies[5]. The commonest site for these tumours was the CBD (40.1%).

Primary cystic duct carcinoma, on the other hand is extremely rare especially if the strict criteria of Farrar are implemented. It amounts to 2.6-12.6% of all extrahepatic bile duct malignancies[6]. Its clinical presentation is nonspecific and does not differ much from that of the more common calcular biliary disease. Twenty-three cases have been reported in the literature so far, 81% presented with right upper quadrant abdominal pain, 41% with abdominal mass and very rarely (four cases) with obstructive jaundice[2-4]. In none of these cases was a preoperative diagnosis made. The tumour was either discovered at laparotomy or on histopathological examination of the specimen[3].

The case presented here is the 24th in the literature, it has some unique features:

(1) it is the first case in the literature to be diagnosed preoperatively.

(2) it presented as a case of obstructive jaundice, which is a rare presentation of this disease.

(3) the obstructive jaundice and the PTC findings mimicked Mirizzi syndrome. Only one case of primary cystic duct carcinoma presented in this way[4].

Once a cystic duct malignancy is suspected pre- or peroperatively, the operative procedure of choice is radical en masse excision of the CBD, cystic duct, part or all of the CHD (until an end free of tumour is found by frozen section) and the gallbladder together with all fat, periductal tissues and lymph nodes. Continuity is attained by biliary-enteric anastomosis[7].

The prognosis of primary cystic duct carcinoma is considered to be fairly good[2]. The average survival was reported to be 20.2 months while that of gallbladder carcinoma was only 5.8 months[8] and of other extrahepatic biliary ducts 3.2-11.4 months[9,10]. This is probably due to early presentation, as obstruction of the cystic duct gives rise to early signs and symptoms of gallbladder disease.

Although endoscopic retrograde cholangiopancreatogram and/or percutaneous transhepatic cholangiogram are essential investigations in cases of obstructive jaundice, simple non-invasive ultrasonography by an experienced radiologist may accurately diagnose the case and probably eliminates the need for these invasive and more expensive techniques.

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Hepatic hydrothorax without ascites: a rare form of a common complication

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Keywords: ascites; hepatic hydrothorax; pleural effusion

Introduction

Pleural effusion of abdominal origin occurs in 5% of cirrhotic patients[1], but the rare cases[2-5] reported in patients without ascites have led to pathogenic and diagnostic confusion.

This report proves that hepatic hydrothorax exceptionally complicates the evolution of cirrhotic patients without ascites and deals with the pathogenic mechanisms involved in this entity.

Case report

A 48-year-old man was admitted at the hospital in September 1987 because of dyspnoea which began 2 days earlier. A laparoscopic hepatic biopsy performed in 1982 had disclosed alcoholic cirrhosis. On examination the patient was tachypnoeic and moderately icteric. The heart was normal. Dullness and abolished breath sounds were found over the lower two-thirds of the right hemithorax. There was hepatosplenomegaly, but no evidence of ascites was found.

The haematocrit was 31%, the platelet count was 48 000 and the white cell count was 2900. Prothrombin time was 35% of control value. Total bilirubin was 92 μmol/l (0-17 μmol/l), alkaline phosphatase 147 U/l (30-115 U/l) and serum albumin 29 g/l. Hepatitis B virus serology and alphafetoprotein were negative. An X-ray film of the chest showed a pleural effusion which occupied two-thirds of the right hemithorax. A thoracocentesis yielded 300 cm^3 of clear yellow fluid which contained 19 g/l of total protein and 41 U/l of amylase. Cultures and cytology were negative. A post-tap chest X-ray was normal. An ultrasonographic examination revealed no evidence of ascites. An intraperitoneal injection of 10 mCi of ^{99}Tc-colloid was followed by a positive uptake of radioactivity on the right pleural cavity from the first hour after injection (Figure 1).

Fifteen days later the patient again presented pleural effusion which resolved after 2 weeks' treatment with diuretics. A third effusion resolved spontaneously. In October 1987 the patient developed ascites and 100 mg of spironolactone was prescribed. One week later he was re-admitted with a pleural effusion which obliterated the entire right hemithorax. A chemopleurodesis with talc was

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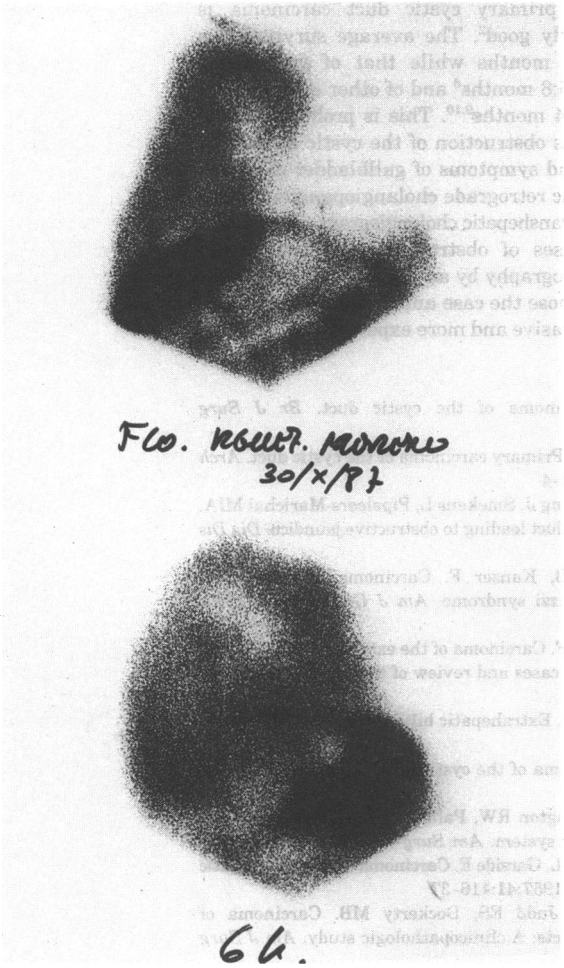

Figure 1.

performed, but 6 days later the patient died in a final phase of hepatic failure.

Discussion

Our results confirm the abdominal origin of hepatic hydrothorax even in patients without sonographic evidence of ascites[6] and serve to suggest the conditions in which spontaneous or diuretic-induced reabsorption occur and the mechanisms by which hydrothorax can be associated with ascites or not, even in the same patient.

The passing of ascites towards the thorax is determined by the sum of the negative pleural pressure (NPP) and the positive abdominal pressure (PAP). When the pleural filling pressure (PFP) equilibrates the former forces (NPP+PAP) the effusion remains stable, but a flow of ascites persists only to replace the liquid which is reabsorbed in the pleural serosa. However the characteristics of this equilibrium vary according to the mechanism of production of the pleural defect. If the hole develops when there is ascites, or is even due to it, the PAP will be very high and, given the limited capacity of the pleural cavity, the persistence of hydrothorax and ascites is logical. Instead, in those patients without ascites a diaphragmatic defect must exist before effusion develops, which permits the passage of liquid as soon as there is a minimal production of ascites. Consequently, in some of these patients, an equilibrium can be established in conditions in which the production of ascites is equal to the pleural capacity of reabsorption, so that ascites will not accumulate. Any decrease in the production of ascites can easily make it smaller than the pleural capacity of reabsorption and the effusion would finish being reabsorbed, as happened in the patient reported here. On the contrary any increase in the production of ascites will lead first to its accumulation and later to its decrease or disappearance if the increase in the PAP forces a net passage of ascites into the thorax, reaching a new equilibrium at a higher level of PFP and a greater hydrothorax, as was observed finally in our patient.

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Rare type of visceral myopathy mimicking anorexia nervosa

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Keywords: small bowel; genetic disease; smooth muscle degeneration

We report a case of visceral myopathy involving small bowel without obstructive symptoms or radiologic intestinal dilatation.

This rare type of visceral myopathy acquired by autosomal recessive transmission is characterized by small bowel involvement without clinical obstruction or radiological dilatation.

Case report

A 19-year-old woman who had been treated for anorexia nervosa for two years was admitted with severe cachexia (height 1.52 m, weight 26 kg), oedema and ascites. She complained of chronic diarrhoea (6 stools/day), abdominal pain and vomiting, but she had no signs of obstruction. Examination threw doubt on the initial diagnosis of anorexia nervosa and led to her having extensive digestive tract investigations culminating in laparotomy.

A plain film of the abdomen did not reveal any bowel obstruction. A barium meal showed a hiatus hernia and a normal oesophagus and duodenum. Oesophago-gastro-duodenal endoscopy confirmed the hiatal hernia and was used to provide proximal small bowel biopsies. Colonoscopy was negative and intubation of the terminal ileal loop was unsuccessful. However, barium X-rays of the small bowel were abnormal showing a diffuse enteropathy involving the entire small bowel (especially its middle part). There was mild thickening of intestinal wall, moderate contraction of

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